in comparison to PMMA, silicon, and hydrogel lenses.[3] This might be relevant in our patient's left eye, as he had a three-piece lens. However, as previous reports have included single-piece PMMA,[4,6] three-piece PMMA,[5] and acrylic[5] IOLs, no definite conclusion can be drawn. These factors are unlikely to be relevant in his right eye.

The posterior luxation of the PCIOL in our patient's right eye raises the possibility of a bilateral involvement, although it could be a result of previous surgery. He had bilateral developmental cataracts and a retinal detachment in one eye, which are common in Marfan's syndrome and suggest a multifactorial aetiology. Although our patient had no family history, he had some features suggestive of Marfan's syndrome—tall habitus, arm span/height ratio of 1.04, upper/lower segment ratio of 0.71, reduced keratometric power, and increased axial length. Spontaneous luxation of an IOL has not to our knowledge been reported, as a presenting feature in Marfan's syndrome to date. These features are, however, not sufficient to make a diagnosis of Marfan's and it is debatable, whether this is a *forme fruste* of Marfan's syndrome. Mengesha[10] reported ectopia lentis in the absence of a family history and other signs and called it a *forme fruste* of Marfan's, implying that not all clinical features are present and the diagnosis is not secure. As genetic mutation occurs in 15% of cases, a family history may not always be positive. Although the mechanism of IOL-bag luxation in our case is inconclusive, it is most likely a manifestation of Marfan's syndrome.

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Sir,

Limited Wegener's granulomatosis of the orbit: a case study and review of literature

Wegener's granulomatosis (WG) is a severe, noninfectious, granulomatous, and necrotizing vasculitis that classically affects respiratory tracts and kidneys.[1] Clinical manifestations of WG are nonspecific and indistinguishable from a variety of neoplastic, infectious, and inflammatory diseases. The limited form of WG usually lacks renal involvement, frequently involves the head and neck region, and may not always progress to generalized disease with a resultant better prognosis.[2,3] We report an unusual presentation of limited WG of the orbit showing extensive orbital involvement without systemic or sinonasal tract involvement. The histopathology showed a classical triad of vasculitis, necrosis, and granulomatous inflammation with *multinucleated giant cells*, which are rarely seen from the orbital specimen.[4]

Case report

A 56-year-old male presented to us in July 2003 with complaints of right-sided gradually progressive proptosis (since August 2002), associated diminution of vision (since January 2003), and ptosis (since May 2003).

He did not give a history suggestive of any systemic involvement. He was operated for right-sided chronic dacryocystitis in June 2002. At presentation, the right eye had just perception of light with inaccurate projection in all quadrants, and the corrected visual acuity in the left eye was 20/60. The examination of the right eye revealed 11 mm nonaxial proptosis (inferonasal), total limitation of movements in all directions, moderate ptosis, lagophthalmos, exposure keratopathy, conjunctival congestion, dilated episcleral veins, immature senile cataract, sluggishly reacting pupils with relative afferent pupillary defect, and secondary optic atrophy (Figure 1a). On palpation there was a firm to hard mass surrounding the globe (maximum superotemporally), non-reducible, noncompressible, nonpulsatile, the posterior extent was not reachable, the overlying skin was mobile, the bony margins were palpable and draining lymph nodes were not enlarged. The left eye revealed only immature senile cataract. The ENT and systemic examination did not reveal any abnormality. The complete blood count was within normal limits, except ESR (60 mm in the first hour). Blood sugar, blood urea, serum creatinine, serum electrolyres, thyroid function tests, urine examination, and abdominal ultrasonography were all within normal limits. Bilateral lung fields were clear on chest X-ray and there was no hilar lymphadenopathy. CT and MRI scan of the orbit were abnormal (Figure 1b,c). cANCA was negative by indirect immunofluorescence. The wedge biopsy taken from the mass was negative for acid-fast bacilli (Zeihl Neilsson stain) and fungal hyphae (PAS Stain). The bacterial culture was sterile, and histopathology showed fibroadipose tissue with acute on chronic inflammation, microabscess formation, and few eosinophils. The patient was given systemic steroids under antibiotic cover. In August 2003, the patient had a blind eye with progressively increasing proptosis necessitating repeated tarsorraphies every week. There was no response to therapy, increased exposure keratopathy, histopathology was inconclusive and mass was now overhanging the orbital margins. The orbital exenteration was carried out and histopathology showed classical triad vasculitis, necrosis, and granulomatous inflammation with multinucleated giant cells, diagnostic of WG (Figure 2).

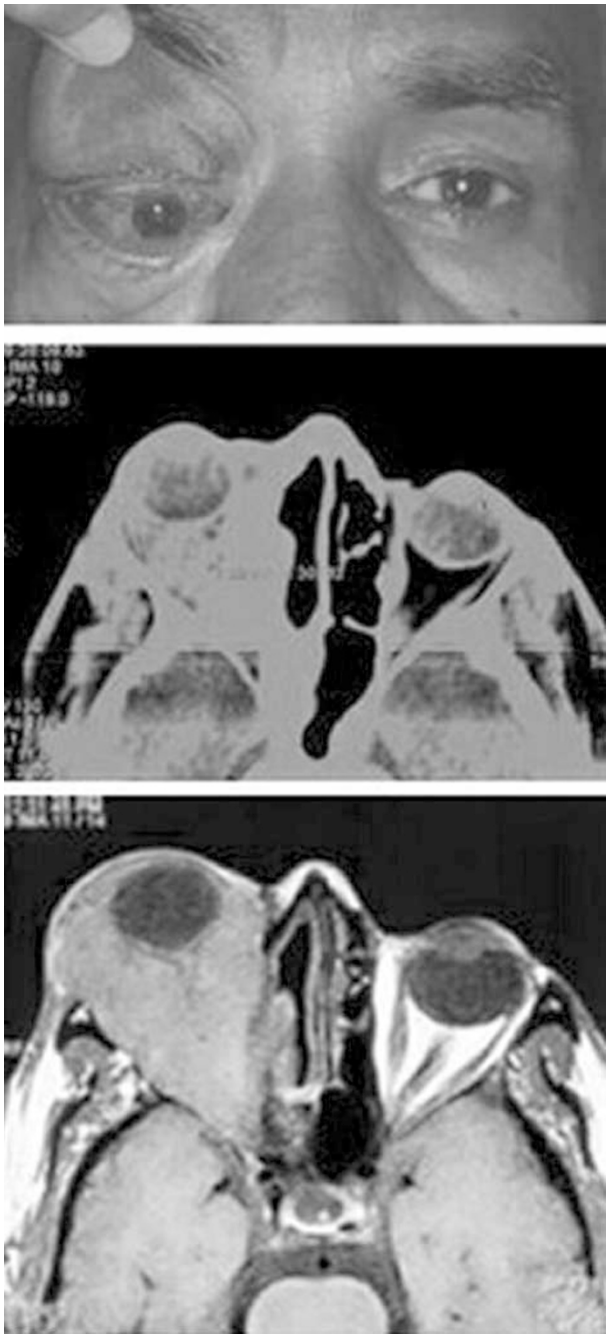

Figure 1 (**a**) A 56-year-old male at presentation showed nonaxial proptosis (inferonasal), moderate ptosis, lagophthalmos, exposure keratopathy, conjunctival congestion, dilated episcleral veins, and immature senile cataract. (**b**) CT scan showing soft-tissue mass occupying both intra- and extraconal spaces of right orbit, with homogenous contrast enhancement and erosion of medial orbital wall. (**c**) MRI scan showing soft-tissue mass occupying both intra- and extraconal spaces of right orbit with obliteration of outline of extraocular muscles and optic nerve complex. There was no obvious bony destruction or intracranial extension.

Comment

In 7% patients of generalized WG, proptosis is the initial presentation. Ocular/orbital involvement is ultimately seen in 50% cases of generalized WG.[5] Orbital involvement in WG occurs either due to contiguous spread from the sinuses or due to primary involvement (small vessel vasculitis). The common ocular features

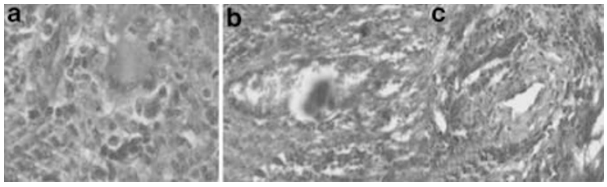

Figure 2 Histopathology showing a classical triad of Wegener's granulomatosis: (**a**) granulomatous inflammation with multinucleated giant cell, (**b**) areas of necrosis, and (**c**) vasculitis.

include[6] proptosis (13%), scleritis and episcleritis (11%), peripheral corneal ulceration (8%), nasolacrimal duct obstruction (7%), optic nerve vasculitis (6%), retinal artery occlusion (5%), conjunctivitis (4%), and uveitis (3%).

The diagnosis of orbital WG is not straightforward because of the confusion with other entities that may present with similar features.[5] When clinical and serological findings are inconclusive as was in our case, biopsy remains indispensable. The histopathologic triad of granulomatous inflammation, tissue necrosis, and vasculitis is seen in not more than 50% of orbital biopsies.[4] The presence of a classical triad is virtually diagnostic, but absence does not exclude the diagnosis of WG.[7] Multinucleated giant cells seen in our case rarely accompany the granulomatous inflammation in orbital biopsies of WG. Fine-needle aspiration cytology has a limited role in the diagnosis of WG because histological features are difficult to detect by cytological methods.

ANCA is not recommended to be used alone in place of a biopsy because ANCA is positive in only 67% cases of limited WG.[8,9] Positive ANCA may help in establishing the diagnosis in cases in which typical pathological features are lacking,[10] and it has a value in following disease activity.[11] In retrospective analysis of 15 patients of limited orbital WG over a period of 23 years, only one patient had severe orbital involvement needing orbital exenteration, whereas orbitotomy was carried out in eight patients, external ethmoidectomy in two patients, and endoscopic transnasal exploration in three patients.[12] Limited WG of the orbit is a rare disease, but with overall good prognosis. It should be kept in the differential diagnosis of patient presenting with proptosis in the fourth or fifth decade.

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